

# Synovial chondromatosis of the distal radio-ulnar joint

David Botros, BS<sup>a,b</sup>, Ken Ford, BS<sup>c</sup>, Brendan Holderread, BS<sup>c</sup>, Al Mollabashy, MD<sup>c</sup>, and James Rizkalla, MD<sup>b,c</sup>

<sup>a</sup>Department of Neurological Surgery, The Johns Hopkins University School of Medicine, Baltimore, Maryland; <sup>b</sup>Department of Orthopedic Surgery, Coptic Medical Association of North America Research Institute, Dallas, Texas; <sup>c</sup>Department of Orthopedic Surgery, Baylor University Medical Center, Dallas, Texas

## ABSTRACT

Synovial chondromatosis (SC) is a benign metaplastic proliferation of cartilaginous nodules within the synovial membrane. Primary SC, though a rare monoarticular disease, significantly impacts patients' functional and pain-related outcomes. We outline the case of a 52-year-old man who presented with a large mass on the volar-ulnar aspect of his left wrist. Biopsy and workup revealed SC of the distal radio-ulnar joint. Though most cases of SC can be managed with arthroscopic or intralesional resection of the mass, a subset of extremely aggressive cases of SC may ultimately fail intralesional resection. Patients must be counseled about the possibility of amputation as an ultimate treatment option for the resolution of their pain and symptoms.

**KEYWORDS** Amputation; mass; orthopedic oncology; resection; synovial chondromatosis; tumor

Synovial chondromatosis (SC) is a benign metaplastic proliferation of cartilaginous nodules within the synovial membrane, often presenting with classically described “loose bodies” occupying the joint space.<sup>1,2</sup> SC occurs in approximately 1.8 per 1 million individuals.<sup>3</sup> Though in theory any articular space is at risk of SC, the literature suggests a predilection to the knee and hip, with nearly 90% of all cases of SC occurring at these sites.<sup>4</sup> This article describes an incredibly rare case of SC of the distal radio-ulnar joint (DRUJ) that was more aggressive than described in previous case reports. The aggressiveness and local recurrence of this lesion ultimately required amputation as the definitive treatment.

## CASE REPORT

A 52-year-old right hand-dominant man presented to the outpatient setting of an orthopedic oncologist with a large soft tissue mass occupying much of his left wrist (*Figure 1a, 1b*). Biopsy of the lesion was consistent with florid SC. Magnetic resonance imaging (MRI) revealed a large, multiloculated cystic mass in the distal forearm and wrist, centered in the flexor compartment, radially displacing and flattening the flexor and extensor musculature and vascular structures. With a confirmed histological diagnosis of SC and an overall tumor size of 10.2 × 11.7 × 9.9 cm (*Figure 1c, 1d*), options for treatment included trans-forearm amputation vs an attempted intralesional resection

of the underlying mass with preservation of the skeleton where feasible. These options were discussed with a multidisciplinary tumor board, as well as the patient, who ultimately decided on intralesional resection and attempted limb salvage. After aggressive resection, including the distal 10 cm of the ulna, the tumor returned 7 months later. A subsequent attempt at limb salvage was pursued for a second intralesional resection. Nonetheless, the tumor's aggressiveness persisted, returning 6 months after the second surgery with additional lesions and pain. At this time, it was discussed with the patient that he would likely benefit most from trans-forearm amputation, because the limb was not functional and continued to be a source of pain. The patient consented, underwent the procedure (*Figure 1e*), and was transitioned appropriately to an upper-extremity prosthesis for utilization in his activities of daily living.

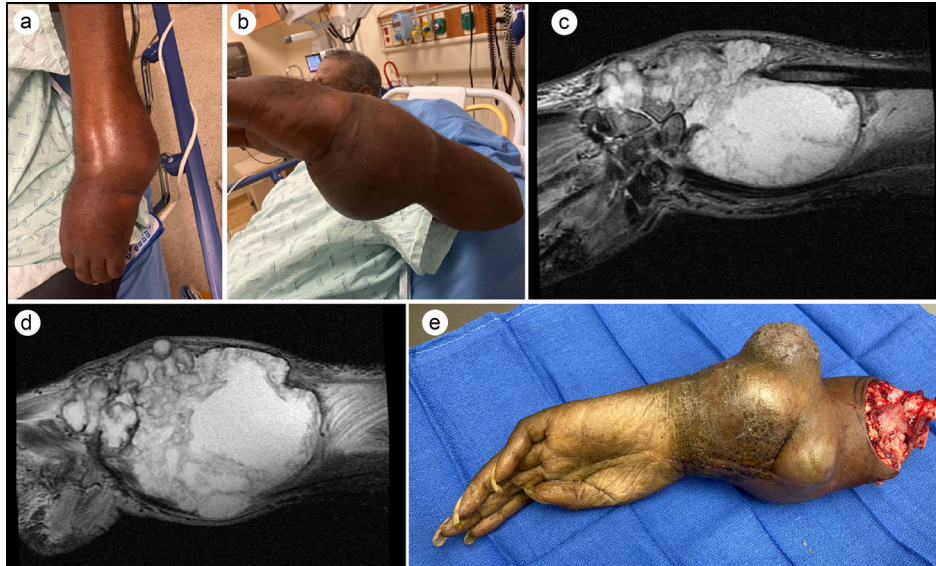
## DISCUSSION

The exact etiology of SC is unclear, though it is commonly thought to involve metaplasia of the pluripotent cells of the synovium to chondrocytes. Limb amputation is usually reserved for the rare occurrence of SC progression to chondrosarcoma. This malignant transformation is rare, with less than 40 cases of histologically confirmed transformation existing in the literature. Rapid recurrence of benign SC was documented preceding malignant transformation in many cases.<sup>5,6</sup>

**Corresponding author:** James Rizkalla, MD, Department of Orthopedic Surgery, Baylor University Medical Center, 3500 Gaston Ave., Dallas, TX 75246 (e-mail: [jrizkall@gmail.com](mailto:jrizkall@gmail.com))

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**Figure 1.** (a, b) Clinical photographs demonstrating a large mass around the left wrist. (c, d) Coronal STIR MRI sequences revealing a large disruptive soft tissue mass primarily on the volar aspect of the distal radio-ulnar joint, with a large centrally necrotic lesion and disruption of osseous structures. (e) Clinical image of amputation stump after definitive surgery showing aggressive regrowth of soft tissue masses around the distal radio-ulnar joint.

Due to its variable presentation and symptomology, diagnostic criteria for SC have not been formally defined. Arthroscopic removal of the cartilaginous loose bodies with partial or full synovectomy is usually preferred for symptomatic SC patients, because it shortens recovery time and postsurgical complications.<sup>7–12</sup> Open surgery, however, may be superior for smaller, less accessible joints (such as SC of the wrist)<sup>1,13–15</sup> or with highly recurrent disease<sup>16</sup> where complete synovectomy is desired. Rates of postsurgical SC recurrence range from 7.1% in the hip<sup>17</sup> to 17% in the wrist.<sup>1</sup> In addition, recurrence often results from incomplete removal of the synovial membrane<sup>18,19</sup> with subsequent reactivation of the three phases of disease following surgery. With such a wide range for recurrence, our report of multiple recurrences within 13 months presents an uncommonly aggressive form of SC.

Though most SC can be managed with arthroscopic or intralesional resections of the mass, there remains a subset of extremely aggressive SC that may ultimately fail intralesional resection. Given the difficulty of ensuring proper resection at the DRUJ, patients must be counseled of the possibility of amputation as an ultimate treatment option for the resolution of their pain and symptoms.

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